

## Giant Cell Arteritis: A Report of Two Cases from Ethiopia

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### Abstract

**Background:** Giant cell arteritis is a systemic inflammatory vasculitis of unknown etiology that occurs in older persons and can result in a wide variety of systemic, neurologic, and ophthalmologic complications. Giant cell arteritis is a granulomatous necrotizing arteritis with a predilection for large and medium sized arteries. It is considered to be uncommon in the black race with very few reports from Africa. We report two 73 and 74 year old Ethiopian male patients who presented with sudden onset unilateral visual loss that was associated with severe throbbing headache on the affected eye side. Temporal artery biopsy showed characteristic of Giant cell arteritis. Both of our patients were treated with oral prednisolone with no further severe complications from the disease on subsequent follow up for one year. One of our patients developed steroid related complication (high blood sugar) hence monitoring and tapering the dose as soon as possible is of crucial value. The delayed presentation of both the patients has contributed for the poor visual recovery after the treatment.

**Conclusion:** The two cases that we reported underscored that any new onset headache in an elderly patient should prompt a thorough evaluation for the possibility of GCA regardless of race. Heightened clinical awareness of the possibility of temporal arteritis in black patients should lead to earlier diagnosis and initiation of immunosuppressive therapy.

**Setting:** University of Gondar Hospital-a tertiary eye care and training center

**Keywords:** Case report; Giant cell arteritis; Anterior arteritic ischemic optic neuropathy; Black race; Ethiopia

**Abbreviations** GCA: Giant Cell Arteritis; ESR: Erythrocyte sedimentation rate

### Introduction

Giant cell arteritis (GCA), also known as temporal arteritis, is the most common form of systemic necrotizing vasculitis in individuals who are over 50 years of age [1,2].

Although GCA affects people in all cultural and racial groups, it has been reported as particularly common in patients with Scandinavian background the incidence being more than 20 per 100,000 populations [3]. In the USA, the annual incidence ranges from 0.5 to 25 per 100,000 people aged 50 years or older and it is uncommon in those with African descent [4]. The single greatest risk factor for GCA in its various forms is advancing age with mean age of presentation being 72 years [5]. Women are affected four times more commonly than men [6].

Patients commonly present with headache, non-specific symptoms (such as fever, weight loss, night sweating, depression and malaise), polymyalgia rheumatica characterized by pain and stiffness in the proximal muscle group, jaw claudication and scalp tenderness [7,8]. But, the most severe preventable complication of GCA and the one of great concern to those diagnosing and treating the condition is

blindness due to anterior arteritic ischemic optic neuropathy usually caused by posterior ciliary artery occlusion; the clinical picture being described as sudden painless loss of vision [9]. And other ophthalmic presentations include diplopia, amaurosis fugax, ptosis, nystagmus, cranial nerve palsies and internuclear ophthalmoplegia [10]. Visual manifestations may be observed in about 20%-30% of patients, with permanent vision loss in 12%-15% [7,8,11]. Although visual manifestations may be intermittent initially, visual deficit is usually irreversible once established [12].

The diagnosis of Giant cell arteritis is usually made by high index of clinical suspicion and some supportive investigations. Erythrocyte sedimentation rate (ESR) and C-reactive proteins (CRP) get elevated and these tests are both sensitive and useful in treatment response follow-up [13-15]. Thrombocytopenia and anemia may also be suggestive [16,17]. Anemia was more commonly observed in patients without severe ischemic manifestations and ESR greater than 100 mm/h exhibited more commonly constitutional syndrome and associated with less visual ischemic events [18-20]. But, the gold standard diagnostic modality for GCA is temporal artery biopsy and the histopathologic findings shows transmural inflammatory infiltrates with numerous multinucleated giant cells, the vascular lumen get narrowed by concentric intimal hyperplasia [21,22]. According to the American college of rheumatology classification criteria for GCA, the diagnosis of Giant Cell Arteritis is made if patients fulfill three out of the five criteria (Table 1) [23].

1.	Age at onset of disease 50 years and older
2.	New onset headache
3.	Temporal artery tenderness or reduced pulsation
4.	Elevated ESR >50 mm/hr.
5.	Abnormal biopsy results: vasculitis characterized by a predominance of mono nuclear cell infiltration or granulomatous inflammation, usually with multinucleated giant cells.

**Table 1:** The American college of rheumatology classification criteria for GCA.

For purposes of classification, a patient shall be said to have Giant Cell (Temporal) Arteritis if at least three of these five criteria are present. (93.5% sensitivity and 91.2% specificity)

Prompt treatment with high-dose steroids is critical to reduce the likelihood of neuro-ophthalmic complications [24]. It is one of the true ocular emergencies which warrant immediate treatment in order to prevent vision loss [8,25]. The symptoms of GCA should respond

rapidly to high dose glucocorticosteroid treatment, followed by resolution of the inflammatory response [26]. In recurrent or resistant GCA, methotrexate or other immunosuppressive (e.g. azathioprine or leflunomide) may be used as adjuvant therapy [27].

We report our experience with two cases of GCA, who were diagnosed and treated at our center (Table 2).

Characteristic	Case 1	Case 2
Age	74	73
Sex	Male	Male
Ethnicity	Amhara	Amhara
Duration of symptoms (days)	3	4
Headache intensity	Severe	Severe
PMR	Present	Present
Constitutional symptoms	Present	Present
Visual loss	Yes	Yes
Jaw claudication	No	Yes
Temporal artery tenderness	Present	Present
ESR (mm/ hour)	95	120
Hemoglobin (gm %)	12.6	14
Platelet count (cells/ mm <sup>3</sup> )	210,500	429, 000
Temporal Artery Biopsy	Giant cells and mixed chronic inflammatory cells	Multinucleated giant cells and intimal thickening
Treatment	Oral Prednisolone	Oral Prednisolone
Response to treatment	Improvement of the headache and constitutional symptoms, normalizing of ESR but no visual recovery No further severe complications from the disease	Improvement of headache and constitutional symptoms, normalizing of ESR but no visual recovery No further severe complications from the disease
Treatment side effect	High blood sugar	None
Follow up duration	One year	One year

PMR: Polymyalgia Rheumatica; ESR: Erythrocyte Sedimentation Rate

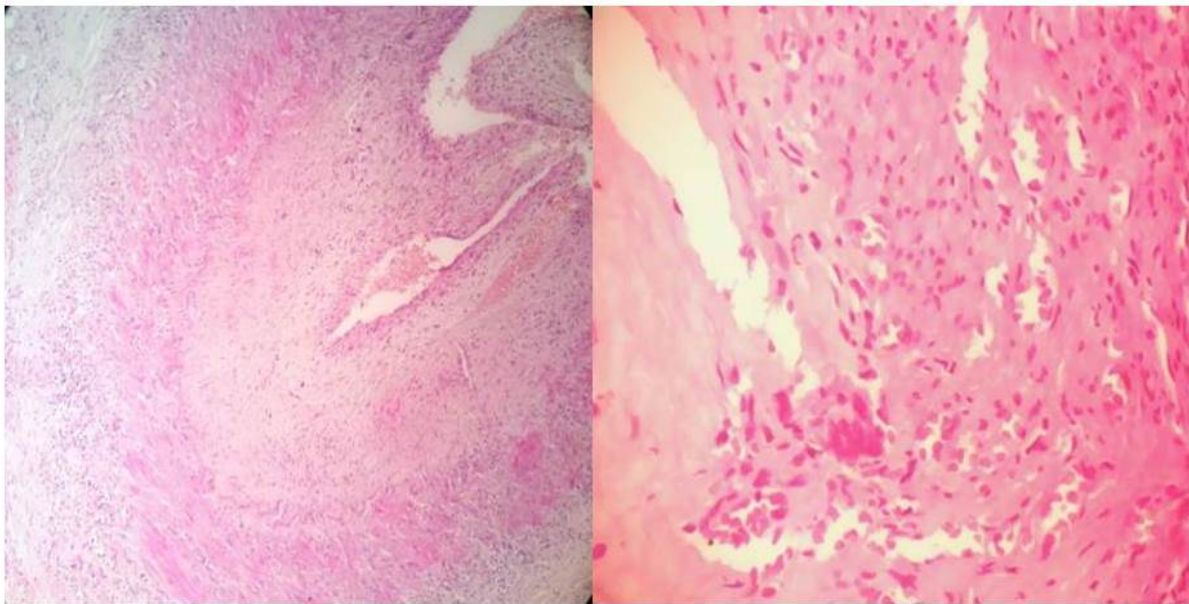
**Table 2:** Summary of clinical findings in the two Ethiopian cases of giant cell arteritis

## Case Report

### Case one

A 74 year old male patient presented with sudden loss of vision of the left eye of 3 days duration. He had severe throbbing headache of the left side that was associated with low grade fever, malaise, loss of appetite and myalgia of the same duration. He had undergone cataract surgery in the right eye 4 months back in our center with good visual outcome. He had no self or family history of diabetes mellitus, hypertension, renal or cardiac disease. Physical findings included left side temple tenderness with cord like temporal artery on palpation. His presenting visual acuity was 6/18 in the right eye and perception of light only in the temporal quadrant of the left eye. Intra-ocular pressure was 22 and 17 mmHg in the right and left eye respectively. He had a relative afferent pupillary defect in the left eye with bilateral heavy pseudo exfoliation on the pupillary margin. A posterior chamber intra-ocular lens was *in situ* in the right eye whereas he had early nuclear and posterior sub capsular cataract in the left. His optic disc was pink in the right but had diffuse pallor with edema in the left.

His complete blood count was within the normal range while his ESR was 95 mm in the first hour. Based on the findings, we made a presumptive diagnosis of left side temporal giant cell arteritis with arteritic anterior ischemic optic neuropathy of the left eye, and he was put on oral prednisolone 1 mg/Kg daily. Left temporal artery biopsy was done 5 days after initiation of steroid treatment and the result came up with mild lymphocytic infiltrates and very few multinucleated giant cells (Figure 1). Two weeks after the initiation of treatment, the patient had improvement of the headache and other constitutional symptoms but no visual recovery in the left eye. At his 4th week of follow up, he presented with generalized malaise and pain over the lower extremities with raised, tender, erythematous lesions on both legs. There was no change in visual acuity of both eyes. His fasting blood sugar was 435 mg/dl and complete blood cell count showed leukocytosis with neutrophil predominance. His ESR was 75 mm in the first hour. With the diagnosis of diabetes mellitus with cellulitis of both legs, he was linked to the internal medicine department of our center where he was successfully treated with the control of both the blood sugar level and his infection.

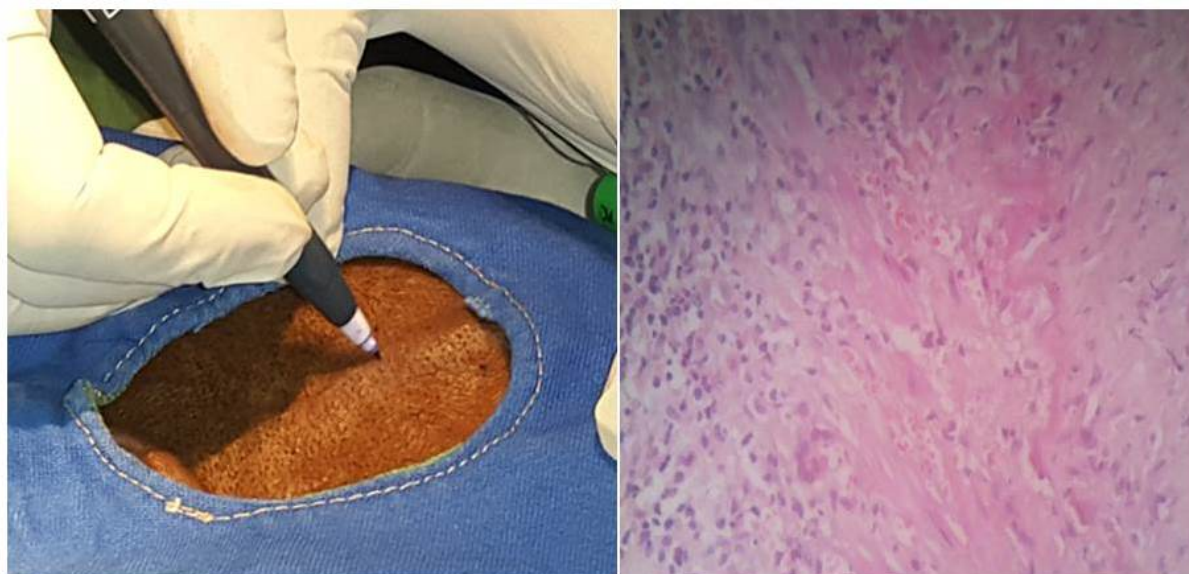


**Figure 1:** Giant cell arteritis of temporal artery, showing giant cells and mixed chronic inflammatory cells in the media of the vessel.

### Case two

A 73 year old male, retired farmer, presented with sudden loss of vision of the right eye since 4 days prior to presentation that was associated with severe throbbing headache of the right side. He also complained low grade fever with loss of appetite, jaw pain that worsen with chewing and muscle ache. He had cataract surgery in the right eye 6 years back and was enjoying good vision, but he had a progressive painless reduction of vision in the left eye with total loss of vision in the same eye since a year back. He had no self or family history of diabetes mellitus, hypertension, renal or cardiac disease. The objective findings included right side temple tenderness with cord like temporal artery on palpation with weak pulse. Visual acuity at presentation was only perception of light in the right eye and no perception of light in left. Intra-ocular pressure was 19 mmHg and 32 mmHg in the right

and left eye respectively. Other ocular findings included pseudo exfoliation at pupillary margins of both eyes; a posterior chamber intra-ocular lens was *in situ* in his right eye while he had early posterior sub capsular and nuclear cataract in the left eye. The optic nerve head was edematous in the right and totally cupped with no neuroretinal rim in the left eye. Complete blood count was remarkable for thrombocytosis of 429,000 cells/mm<sup>3</sup>. Erythrocyte sedimentation rate was 120 mm in the first hour. With the presumptive diagnosis of right giant cell temporal arteritis with arteritic anterior ischemic optic neuropathy of the right eye and absolute pseudoxfoliative glaucoma in the left eye, he was started on oral Prednisolone 1 mg/kg daily. His temporal artery biopsy result showed multinucleated giant cells with intimal thickening (Figure 2). On subsequent follow up, he had improvement of headache and constitutional symptoms, normalizing of erythrocyte sedimentation rate but no improvement of his vision.



**Figure 2:** Note the thickened right temporal artery. Temporal Artery biopsy, showed medial lymphocytic infiltrates with very few multinucleated giant cells and intimal thickening.

## Discussion

Though traditionally it said that GCA is not common in black race, the sudden loss of vision in one eye associated with new onset headache and temple tenderness, and ESR values higher than 50 mm in the first hour led to the presumptive diagnosis of GCA in our patients. Though literature indicated that women are affected four times more commonly than men [6], both of our cases were males. There was similar observation from the case report of GCA in African American by Gilbert et al. suggesting GCA may not have typical clinical presentation in black race [28].

One of the most severe preventable complications of GCA is blindness due to anterior arteritic ischemic optic neuropathy, which was evident in both of our cases. It is said that significant number of patients with GCA complicate with permanent vision loss and early treatment is of great importance to prevent the same complication in the fellow eye [8,25].

The positive rate for temporal artery biopsy has been reported to range from 75% to 96% [29,30] with characteristic pathological features showing the presence of giant cells, mono nuclear infiltrates or granulomas in association with the different features of necrotizing arteritis, and rarely, fibrinoid necrosis [21,22]. In our cases the biopsy showed multinucleated giant cells infiltration with intimal thickening, which is the commonest pathologic feature of GCA.

Once the diagnosis of GCA is considered, treatment (steroid) should be initiated as early as possible in order to prevent neuro-ophthalmic complications such as vision loss and stroke [31]. If one eye is involved, there is a high risk of bilateral vision loss with further delay or stoppage of treatment [32,33]. The optimum dose and duration of prednisolone is still controversial, it can range from 20 to 80 mg per day and usually last 1-2 years with gradual tapering [24-27,34]. Both of our patients were treated with oral prednisolone with no further severe complications from the disease on subsequent

follow up for one year. One of our patients developed steroid related complication (high blood sugar) hence monitoring and tapering the dose as soon as possible is of crucial value. The delayed presentation of both the patients has contributed for the poor visual recovery after the treatment.

The two cases that we reported underscored that any new onset headache in an elderly patient should prompt a thorough evaluation for the possibility of GCA regardless of race. Heightened clinical awareness of the possibility of temporal arteritis in black patients should lead to earlier diagnosis and initiation of immunosuppressive therapy. Temporal artery biopsy is the gold standard diagnostic tool where a segment of at least 2 cm of the artery will need to be removed, as areas of pathology (“skip lesions”) are often interspersed with areas of normal artery.

## Declaration

Ethics (and consent to participate): Not applicable

## Authors' Contributions

FA: Clinically diagnosed and treated the patients and drafted the manuscript. YM: Collected literature and co-wrote the manuscript. WT: Made the pathologic diagnosis, revised the manuscript. All authors read and approved the final manuscript. Availability of data and materials: Pictures of the pathologic slides will be available.

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